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Intraosseous epidermoid cyst of the skull: case study and radiological imaging considerations

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Abstract

We report an atypical case of an epidermoid cyst associated with a skull defect to emphasize the diagnostic utility of bedside ultrasonography for the evaluation of subcutaneous scalp nodules. A 24-year-old woman presented with what appeared to be a benign cyst on the right parietal scalp. The cyst was first noticed one year prior to presentation and caused only mild irritation. Upon excision of the cyst, a notable calvarial defect was found in the frontoparietal bone and surgical excision was suspended. Head and brain imaging was performed confirming the calvarial defect with mild mass effect on the brain. A neurosurgical consultation was obtained, and the patient underwent craniotomy followed by cranioplasty with titanium plate placement. The histological evaluation confirmed the suspicion of an intraosseous epidermoid cyst. We hope to raise awareness of the potential for intraosseous involvement of otherwise routine scalp nodules and emphasize the utility of bedside ultrasonography as a quick, easy, and benign imaging modality to assist in preoperative evaluation.

Keywords: epidermoid cyst, intraosseous, calvarial defect, ultrasound

Introduction

There are a variety of routinely encountered cysts of the head and neck, most commonly epidermoid cysts (49%), trichilemmal cysts (27%), and dermoid cysts (22%), [1]. Epidermoid cysts are common benign tumors that may be encountered on nearly any location of the skin with a relatively high occurrence on the scalp and face [1]. Their peak incidence is in the third and fourth decade of life. When found within the skull, they are often located in the frontal and parietal bones [2]. Reports of epidermoid cysts causing intraosseous erosion of the calvarium are found in the medical literature; however, their occurrence remains relatively rare. It is important to identify when to utilize such services as diagnostic radiology or neurosurgery to contribute to a successful diagnosis and treatment. We believe the use of ultrasonography would have been an exceptional additional imaging modality and diagnostic adjunct in the following case of a patient with an intraosseous epidermoid cyst of the scalp with an underlying calvarial defect.

Case Synopsis

A 24-year-old woman was referred to our dermatology clinic for evaluation of a cystic nodule on the right parietal scalp. She first noted the area in April of 2012 when she was on a military deployment to Afghanistan. She noted the lesion had not grown significantly over time and was only mildly tender when wearing her Advanced Combat Helmet. Upon presentation, a 2.5-centimeter ovoid, firm nodule was palpated on the right parietal scalp. The nodule had apparent mobility yet was not compressible with light-to-moderate pressure. The overlying skin was of normal thickness, but with a high degree of laxity. She denied any presence of headaches, blurry vision, tinnitus, memory problems, numbness, paresthesias, weakness, difficulty with gait, bowel or bladder dysfunction, nausea, or vomiting. Differential diagnoses of the nodule included trichilemmal cyst, epidermoid cyst, and subgaleal lipoma.

After preoperative evaluation, surgical removal was attempted. Upon excision, a firm cyst was identified that easily ruptured, releasing a cheesy, gray-colored material. Unexpectedly, an oval calvarial defect approximately 2.5×2.0cm in size became apparent. The excision was suspended, and the overlying skin was closed. Follow-on computed tomography (CT) and magnetic resonance imaging (MRI) evaluations were arranged and neurosurgery consultation was obtained.

CT scan demonstrated a 2.5×1.8cm calvarial defect over the right frontoparietal region that showed erosion of both the inner and outer table of the calvarium. The bony margins adjacent to the lesion appeared smooth and demonstrated remodeling consistent with a chronic process. Subsequent MRI similarly showed right parietal calvarium erosion of both the inner and outer tables, demonstrating a light bulb bright appearance on diffusion-weighted imaging and characteristics consistent with an epidermoid cyst (**Figure 1**). The lesion demonstrated a mild mass effect on the adjacent brain parenchyma; however, no signal abnormalities or pathologic post-contrast enhancement were noted. There was smooth remodeling of the calvarial margins abutting the lesion consistent with a benign, slow-growing process. Ventricles and cisterns appeared of normal size, shape, and configuration. The underlying brain parenchyma appeared unremarkable, with no other mass, bleed, acute infarct, or midline shift. The patient was referred for a neurosurgical consultation where she underwent right-sided craniotomy followed by cranioplasty with titanium plate placement. Postoperative pathology results confirmed an intraosseous epidermoid cyst (**Figure 2**). At the time of this publication, she has had no further complications and has returned to normal activities.

In our patient, the differential diagnosis included trichilemmal cyst, epidermoid cyst, or subgaleal lipoma (in descending order), owing to the location, size, shape, and consistency of the lesion. Given the relatively large size (2.5cm) — along with the possibility of the lesion being a subgaleal lipoma —

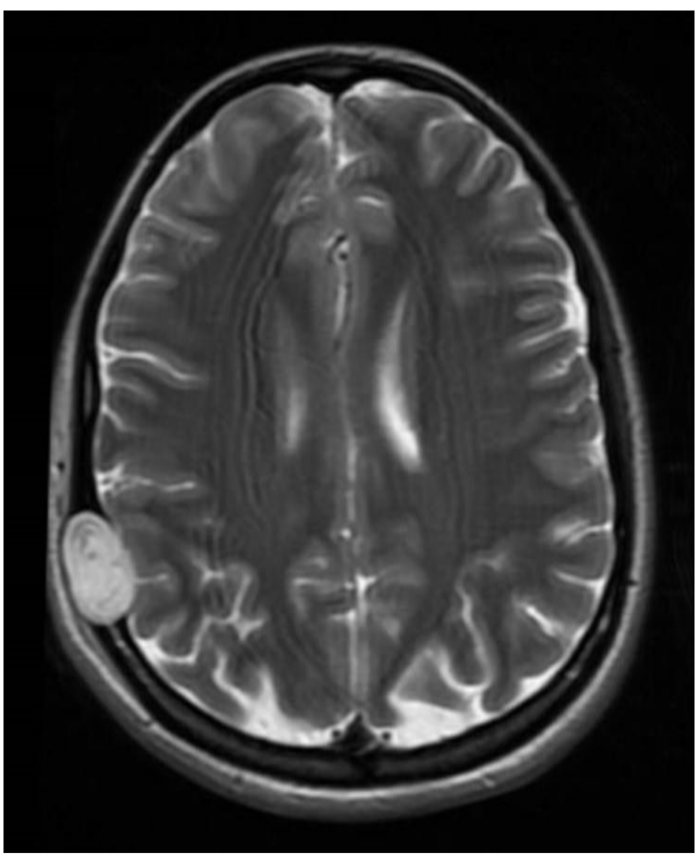

Figure 1. *T2-weighted magnetic resonance image showing a 2.5cm lesion of the right parietal calvarium.*

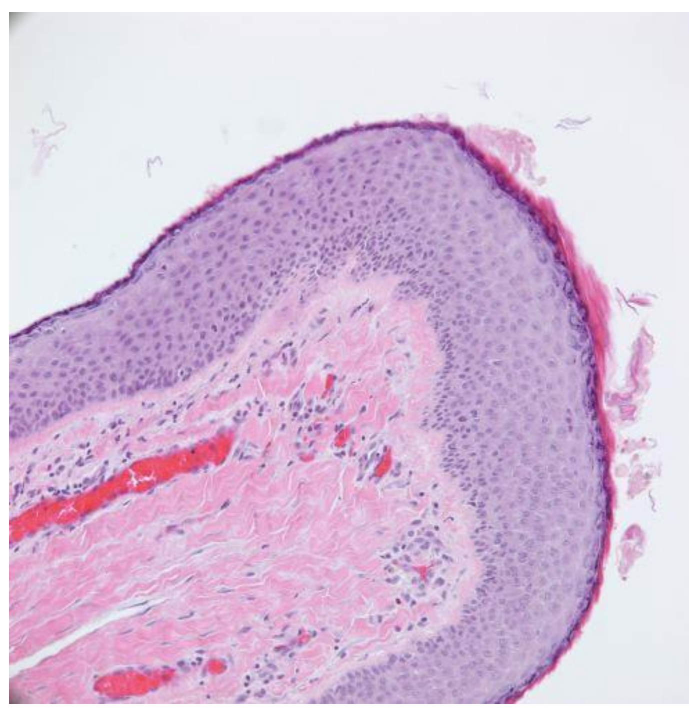

Figure 2. *Skin Biopsy showing an epidermoid cyst with a stratified squamous epithelial wall surrounding inner keratinous material. H&E, 20×.*

we expected to find a limited mobility of the nodule on preoperative examination. These characteristics along with the lack of obvious bony defect on palpation of the circumferential margins indicated the low probability of calvarial invasion. It was not until operative exploration and subsequent cyst rupture that the presence of intraosseous involvement became apparent. In retrospect, this situation would have been an optimal opportunity for the use of bedside ultrasound examination and would have allowed us to surgically triage this patient more appropriately.

Case Discussion

Intraosseous epidermoid cysts account for approximately 1% of all intracranial tumors [3]. As such, we found relatively few mentions of comparable cases during our review of the medical literature. We did, however, review a case with similar calvarial involvement that was believed to be a result of long-term pressure of the cyst creating an erosion of bone known as scalloping. Scalloping is considered to be a benign radiographic feature that is typically absent in malignant processes [4]. Like our case, the authors noted no preceding history of trauma in the patient and an otherwise benign presentation.

In our case, the presence of smooth calvarial margins and evidence of chronic remodeling noted on both MRI and CT led us to believe the cyst had grown slowly over time. It was difficult to determine whether this epidermoid cyst was acquired or rather the long-term manifestation of a congenital abnormality, as epidermoid cysts can arise from both circumstances [5, 6]. Congenital epidermoid cysts are presumed to result from epidermal rests of cells becoming entrapped during development, which slowly manifests over time into palpable, firm, non-tender nodules [5, 6]. Acquired epidermoid cysts are often caused by trauma or some iatrogenic cause bringing epidermal tissue structures deeper into the dermis where they become entrapped [5, 6]. In our case, the patient recalled no unique mechanism of injury or trauma to the area, nor was there a history of any prior surgeries to support an iatrogenic cause. These factors, in addition to our patient's young age, increased the likelihood that the cyst may have been congenital.

The key point to our case was determining the necessity of imaging prior to surgical intervention. In the majority of cases, tactile examination alone provides an accurate preoperative diagnosis. In our case, there was apparent mobility to the nodule. This perception was related to the significant laxity of the overlying skin and possible movement of the nodule within the calvarial defect. However, the thickness of the scalp limited the ability to detect cortical changes on the peripheral margins of the cyst. These limitations on tactile examination are precisely when radiological imaging would prove useful.

If imaging was to be obtained, however, further questions arose: Which modality would contribute the highest diagnostic yield? Which modality would be best to determine the extent of the mass into the adjacent calvarium? And how would it ultimately alter our treatment? To answer these questions, we reviewed existing medical literature, focusing on cases of diagnostic imaging used in the evaluation of epidermoid cysts.

It is common practice for midline scalp lesions and early onset or questionable non-midline cranial suture defects in adults to receive radiologic imaging, given the risk for potential intracranial extension [7]. Plain skull x-rays are able to detect lytic lesions such as myelomas, metastases, and Langerhans cell histiocytosis [7-9]. Certain tumors may also be denser than surrounding bone — such as osteomas and meningiomas — and may also be visualized on plain films. In contrast, dermoid and epidermoid tumors (without central connection) and lipomas cannot be seen on standard x-rays [10]. The inability for routine skull X-rays to detect these types of neoplasms is where other imaging techniques prove useful.

CT and MRI are proven modalities when it comes to skull and intracranial imaging. CT is relatively quick and easily demonstrates calvarial erosions, mineralized components of lesions [10], and the underlying brain parenchyma, but involves exposure to ionizing radiation, which may be undesirable in pediatric and young adult populations and thus limits its routine use. MRI is also capable of depicting tissue character and vascularity of various lesions [11]. However, its availability is often limited, costly, and not always well tolerated by patients.

Ultrasound has a long history as a safe, user-friendly imaging modality that has significant value to dermatologic evaluations. Through variations in echogenicity, ultrasound can differentiate a variety of dermatologic conditions and neoplasms. It has been utilized in evaluating the depth of basal cell carcinomas and melanomas and can monitor disease progression of psoriasis and scleroderma over time [12, 13]. It also has demonstrated utility for differentiating inflamed lymph nodes from those harboring metastatic disease [14]. Lastly — and likely the most practical use of ultrasound in dermatology — is its ability to scan soft tissue tumors (lipomas) and dermally-located tumors (epidermoid cysts and dermoid cysts) to provide information regarding size, depth, and proximity to vascular structures.

A retrospective study of 183 patients with benign subcutaneous lesions was performed assessing the diagnostic yield of ultrasound examination in addition to clinical exam [15]. The authors found a statistically significant improvement in diagnostic accuracy when ultrasound was added to clinical examination of epidermoid cysts. Diagnostic accuracy rose to 99.3% when ultrasonography was combined with palpation, as opposed to 93.5% accuracy by palpation alone ($P<0.05$). This evidence suggests that ultrasound is indeed an important diagnostic tool for evaluating subcutaneous nodules.

The use of ultrasound to diagnose bony erosions has been well documented in individuals with rheumatoid arthritis. However, the diagnostic accuracy appears to vary depending on the area of the body in which it is used [16-18]. There are multiple reports of sonography being used as a safe and effective diagnostic tool in pediatric skull fractures, particularly in the emergency setting. A prospective observational study of 767 consecutive pediatric head injury patients was performed comparing the diagnostic accuracy of bedside ultrasound to CT [19]. The study demonstrated 100% sensitivity (95%, CI 88.2%-100%) and 95% specificity (95%, CI 75.0-99.9%) for ultrasound use in diagnosing pediatric skull fractures as compared to CT scan. They cited simplicity of use, minimal operator training, lack of radiation exposure, and high diagnostic yield as key points for utilizing ultrasound over CT scan.

Another prospective cohort study of 212 children and young adults with 348 suspected fractures was performed evaluating the effectiveness of point-of-care ultrasound diagnosis of fractures in various long and non-long bones [20]. Sonographers participating in the study were emergency medicine physicians with varied ultrasound experience who received a one-hour block of instruction prior to participating. CT scans and standard X-rays were used as reference standards for comparison against ultrasound evaluations. Although the sensitivity and specificity varied depending on the type of bone being imaged (e.g. long bones versus non-long bones), accuracy for identifying cortical disruption indicative of skull fractures was 100 percent. The study did not specify the age of the patients with clinically identifiable skull fractures, but it nonetheless shows that clinicians with minimal formal radiological training can easily and accurately diagnose cortical disruptions of skull bones and identify basic skull anatomy using ultrasonography (**Figure 3**). The authors highlighted the increased availability of ultrasound devices, the versatility of use, and accuracy in identifying cortical disruptions as important reasons to consider sonography as an

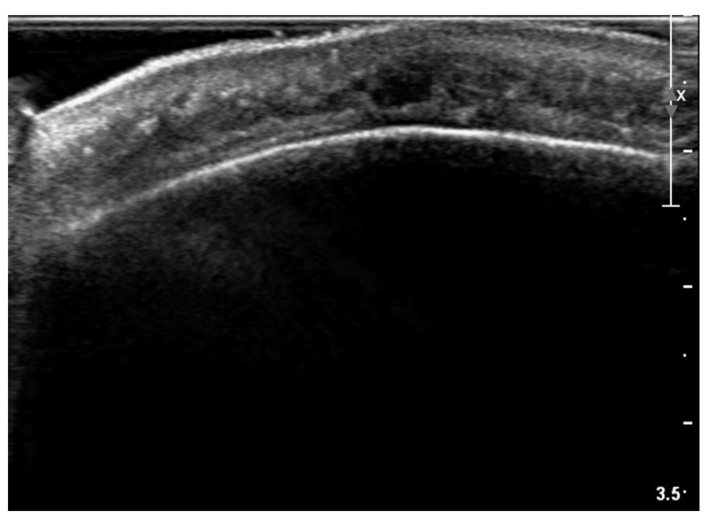

Figure 3. *Ultrasound image showing normal cortical bone and trabecular bone.*

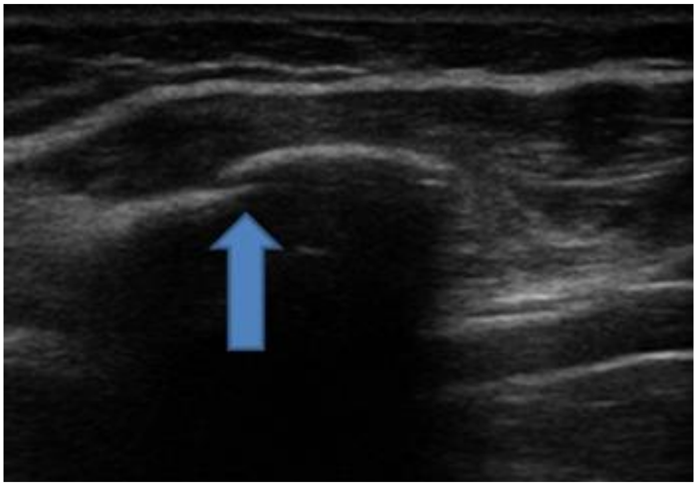

Figure 4. *Ultrasound image showing cortical bone fracture (Courtesy of Giles N. Cattermole, BM BCh).*

initial imaging modality, especially in environments where x-ray or CT are not readily available.

Conclusion

Ultrasound may indeed be an important and useful tool in diagnosing young adult calvarial defects owing to its simplicity, safety, and availability. Moreover, the evidence suggests that diagnosis of cortical fractures (**Figure 4**) can be made with relative ease using ultrasound as a primary imaging modality. The same ultrasound diagnostic techniques can be expanded to include the evaluation of neoplasms in areas at increased risk for intracranial extension, such as the midline scalp or cranial suture areas. Ultrasound may also provide key diagnostic clues to assist the clinician in determining whether to pursue additional imaging studies (such as CT scan or MRI) prior to surgical excision. We believe using ultrasound, especially in young adult patients, should be highly considered as part of a preoperative workup, particularly if there is any question regarding the time of onset, history of trauma, mid-line or cranial suture location, or suspicion of cortical changes. From a practical standpoint, a non-midline scalp neoplasm in a middle-aged or older adult may not be considered cost-effective or high yield but should be considered in circumstances when the history and physical examination dictate.

As the availability of ultrasound devices expands there may be a time soon when more dermatologists will incorporate these devices into clinical practice. Furthermore, the trained eyes and tactile sense of the dermatologist combined with ultrasound examination may lead to unparalleled diagnostic accuracy and patient safety. We hope to raise awareness of this modality to assist in the preoperative evaluation of scalp nodules and sense it may be time to embrace the use of ultrasound more widely within the field of dermatology.

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